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Ectopic Cushing's Syndrome: Importance of Methodical Approach to Hide and Seek

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Cushing's syndrome (CS) is associated with high morbidity and fivefold excess mortality. Ectopic ACTH syndrome accounts for 20% of CS cases. It is crucial to identify and localize the source of extrapituitary ACTH secretion because surgical resection of the primary, often malignant, tumor has a high probability of cure with complete remission in 80% of cases. A 72-year-old female was admitted to a community hospital for lateral chest pain after a fall. She endorsed progressive weight gain, new HTN, emotional lability and worsening cognition over the prior 6 months. ACTH and cortisol levels were elevated. MRI brain failed to identify a pituitary lesion. CT chest showed multiple rib fractures and anterior mediastinal mass which was biopsied. While awaiting pathology results, IPS sampling attempted but aborted due to pulmonary edema necessitating ICU care. She was transferred to our institution for planned TSR of suspected pituitary adenoma. Examination showed Cushingoid features. Labs showed hypokalemia, high cortisol and high ACTH. MRI pituitary showed normal sella. MRI spine showed multilevel vertebral compression fractures. CT chest showed interval growth of mediastinal mass. A 1 mg dexamethasone suppression test failed to suppress ACTH and cortisol, confirming ACTH dependent hypercortisolism. A high dose 8 mg dexamethasone test failed to suppress ACTH and cortisol, a 10 mcg DDAVP stimulation test showed a flat response, with ACTH and cortisol levels remaining comparable before and after DDAVP injection. Dynamic testing therefore consistent with ectopic ACTH dependent hypercortisolism. Gallium-68 DOTATATE PET Scan demonstrated uptake in mediastinal mass suggestive of somatostatin receptor positive neuroendocrine tumor. Biopsy specimens processed at our institution showed cells positive for epithelial markers Cytokeratin and PAX8. Sample showed neuroendocrine differentiation with positive staining for Chromogranin A, synaptophysin and CD 117. Findings consistent with neuroendocrine thymic neoplasm. The patient was started on oral ketoconazole to control hypercortisolism. Surgical resection of tumor was planned with intent to cure CS. Ultimately, due to proximal myopathy and multiple vertebral fractures her performance status was too poor for safe tumor resection. Unfortunately, she died of complications related to CS while attempting preoperative rehabilitation. This case illustrates the importance of a timely, systematic evaluation for ACTH source in ACTH dependent hypercortisolism. Delay in CS diagnosis and localizing correct ACTH source led to missing the window of fitness for curative surgery. If ACTH is elevated, the most common source is pituitary neoplasia, however ectopic ACTH production must be ruled out. Dynamic testing including high dose dexamethasone suppression test, CRH and DDAVP stimulation tests should be used for preliminary localization of ACTH production. IPS sampling is invasive with greater potential for complications, as in the case of our patient. Therefore, it is only indicated if there are conflicting results from different dynamic tests and imaging studies.

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